

## Bovine Spongiform Encephalopathy and Creutzfeldt-Jakob Disease

### What is bovine spongiform encephalopathy?

Bovine spongiform encephalopathy (BSE) is a progressive neurological disorder of cattle that results from infection by an unconventional transmissible agent. It is also called “mad cow disease” because in its advanced stage, cattle stumble around and act strangely. The nature of the transmissible agent is unknown. Currently, the most accepted theory is that it is a modified form of a normal cell surface component known as prion protein.

### What are the symptoms in cattle?

The clinical symptoms of BSE are varied. Most cattle with BSE show a gradual development of symptoms over a period of several weeks or even months, although some can deteriorate very rapidly. Only a small proportion of affected cattle show what would be considered typical “mad cow” signs. Most suspects show several (but not all) of the following symptoms if they are observed closely enough:

- apprehensiveness
- nervousness
- reluctance to cross concrete, turn corners, enter yards, go through doorways or permit milking
- occasional aggression directed at other cattle or humans
- manic kicking when milked
- head shyness, with head held low
- high stepping gait, particularly hind legs
- difficulties in rising
- skin tremors
- loss of condition, weight, or milk yield

### How do the cattle get BSE?

BSE is passed from animal to animal usually by infected feed. Parts of slaughtered animals are often used in feed for other animals.

### What is variant Creutzfeldt-Jakob disease?

Since 1996, strong evidence has accumulated for a causal relationship between BSE and new variant Creutzfeldt-Jakob disease, or just variant CJD (vCJD). vCJD is a rare, fatal brain disorder which causes a rapid, progressive dementia and associated neuromuscular disturbances. There has never been a case of vCJD that did not have a history of exposure within a country where this cattle disease, BSE, was occurring. So currently it is believed that people contract vCJD by consuming BSE-infected cattle products.

### How is vCJD different from CJD?

vCJD should not be confused with the classic form of CJD that is endemic throughout the world. CJD can afflict anyone, but it more often affects people over 60. The median age of vCJD patients is 28 years. Both disorders are invariably fatal brain diseases with unusually long incubation periods measured in years and both are caused by an unconventional transmissible agent. The three apparent means through which CJD may be acquired are (1) sporadically, meaning without apparent cause, (2) the disease can be inherited, or (3) it can be transmitted through infection.

### What are the symptoms of vCJD and CJD and how do they differ from each other and BSE?

All are diseases that affect the brain and the nervous system which causes deterioration of mental function, confusion, possible change in personality, memory loss, tremors, inability to control muscles, and irregular and involuntary contraction of muscles. All also result in holes in the neurons of the brain, giving it a “sponge-like” appearance.

### Is BSE occurring in the United States?

According to the Animal and Plant Health Inspection Service of the U.S. Department of Agriculture, BSE has not

been detected in the United States, despite active surveillance and monitoring efforts since May 1990. Further, to prevent BSE from entering the United States, severe restrictions were placed on the importation of live ruminants (cloven-hooved, cud-chewing animals, including cattle, sheep, bison, buffalo and others) and certain ruminant products from countries where BSE was known to exist. These restrictions were later extended to include importation of ruminants and certain ruminant products from all European countries. There was an outbreak of the disease in the United Kingdom in the late 80s and early 90s. As of 2000, more than 177,500 cases of BSE were confirmed in the U.K. alone. Imports from Canada were suspended in May 2003 also, because of a single case of BSE there.

### **What countries have experienced bovine spongiform encephalopathy?**

Many countries worldwide have seen cases of BSE including the Austria, Belgium, Canada, Czech Republic, Denmark, Finland, France, Germany, Greece, Ireland, Israel, Italy, Japan, Luxembourg, Liechtenstein, the Netherlands, Poland, Portugal, Slovakia, Slovenia, Spain, Switzerland, and the United Kingdom. Not all of them have had cases of vCJD though.

### **Is there any monitoring of the incidence of Creutzfeldt-Jakob disease taking place in the United States?**

Yes. The possibility that BSE can spread to humans has focused increased attention on the desirability of national CJD surveillance. The Centers for Disease Control and Prevention (CDC) monitors the trends and current incidence of CJD in the U.S. by analyzing death data on death certificates with multiple cause of death through the National Center for Health Statistics at the CDC.

### **How would I know if I had vCJD?**

Best estimates now are that the incubation period can be up to ten years, so chances are you wouldn't have any idea exactly where or when you contracted it, but there is no evidence that it occurs in the U.S. First symptoms might be memory problems, inability to control your muscles, or cognitive impairment.

### **Is there a vaccine for CJD or vCJD?**

There is as yet no vaccine, but research is ongoing.

### **Is there a cure for CJD or vCJD?**

At the present time, there is no known effective treatment to arrest or cure CJD or vCJD. The diseases are invariably fatal. The only treatments available focus on easing symptoms and discomfort.

### **What can I do to prevent getting CJD or vCJD?**

To reduce the possible risk of acquiring vCJD from food, travelers to Europe or other areas with indigenous cases of BSE or at possibly increased risk of BSE may wish to consider:

- avoiding beef and beef products altogether or
- selecting beef or beef products, such as solid pieces of muscle meat (versus calf brains or beef products such as burgers and sausages), which might have a reduced opportunity for contamination with tissues that may harbor the BSE agent.

For more information about BSE or vCJD refer to the Centers for Disease Control and Prevention website at [www.cdc.gov](http://www.cdc.gov), the World Health Organization website at [www.who.org](http://www.who.org), or the Food and Drug Administration's website at [www.fda.gov](http://www.fda.gov).